

THE BRITISH JOURNAL
OF
**CHILDREN'S
DISEASES.**

EDITED BY GEORGE CARPENTER, M.D.

REPRINT

PRINTED AND PUBLISHED BY ADLARD & SON,
BARTHOLOMEW CLOSE, LONDON, E.C.

CONGENITAL TROPHIC ŒDEMA.

By OTTO GRÜNBAUM, M.D., D.Sc.

Grünbaum's case

CONGENITAL TROPHIC ŒDEMA.

By OTTO GRÜNBAUM, M.D., D.Sc.*

DURING the last few months three cases of congenital trophic œdema have come under my observation.

CASE 1.—T. R—, a male child, aged 3 months, was brought to the Belgrave Hospital for Children because the mother had noticed that the feet were swollen. The parents are healthy and have a child, aged 2 years, which is to all appearances normal. There is no history of abnormal labour, nor any suggestion that the cause of the deformity is due to maternal impression. I remember seeing a case some years ago which was ascribed by the mother to a fright from an elephant during the period of gestation! The feet alone are affected at the present time; the œdema is symmetrical. There are constrictions at each of the joints of the toes and at the ankles. The child is microcephalic, the circumference of the head being 27·5 centimetres.

CASE 2.—The mother of the second child tells me that her mother had swollen legs "during all her life," but there seems to be some doubt whether the œdema was congenital. The father is a painter, but has never suffered from any signs of lead-poisoning nor from any illness whatever. The mother says that her feet swell towards evening, but are normal in the mornings. There are three children—a boy, aged 6 years, who is normal; a girl, aged 5 years; and a baby, aged 6 months; these two latter are affected with œdema of the feet and legs.

M. H— is an intelligent, well-nourished girl with well-marked œdema, and on her I have had an opportunity of making a few observations. The feet, legs, and lower half of the thighs are swollen, the two sides are symmetrical. That this œdema is con-

* Read before The Society for the Study of Disease in Children, October the 21st, 1904.

siderable may be seen by comparing the measurements of her legs with those of a normal child of the same age and build :

| | M. H. | Normal child. |
|-----------------------------------|----------|---------------|
| Circumference of foot over instep | 17.5 cm. | 13.7 cm. |
| Around ankle | 17.5 cm. | 13.7 cm. |
| Around calf | 20.6 cm. | 20.0 cm. |
| Around knee | 22.7 cm. | 21.2 cm. |

It is evident that the greatest œdema is of the foot and ankle. The toes are very swollen and constrictions mark the joints. The œdema is hard and only pits upon prolonged pressure. The legs are not tender, nor do they ache. Although the mother asserts that the size of the legs is just the same when the child is taken out of bed as when she is put to bed in the evening, nevertheless posture does make some slight difference in the limbs, but the difference in measurement is insignificant in comparison with the alteration in the consistency of the œdema. The legs are considerably harder after having been dependent for some time. The application of a rubber bandage to them diminishes the circumferential measurement by 5 to 7 mm.

The colour of the skin is normal; the temperature of the legs does not suggest any poverty of circulation.

The blood-pressure in the brachial artery is 91 mm. of mercury. This is the maximum, measured by the oblitative method with C. J. Martin's modification of the Riva Rocci sphygmometer.

It is no easy matter to decide whether the skin of the legs is thickened, for the œdema of the subcutaneous tissue leads one to that conclusion upon the first examination; after prolonged massage I was unable to satisfy myself that there was any permanent pathological alteration of the skin.

Sensation to light touch, pressure, heat, cold, and pain was normal.

A detailed examination of the blood did not seem to me to be devoid of interest, since I thought that it might possibly assist me to decide whether the œdema is due to dilated lymphatics or to excess of lymph in the connective-tissue cells. Blood was taken from a finger and from an œdematous foot before and after massage. Care was taken to make a sufficiently deep incision with a small lancet to lead to a free flow of blood without manipulation. The blood from the finger was found to contain

| | |
|-----------------------|-------------------------|
| Red cells | 5,500,000 per cubic mm. |
| White cells | 7600 " " " |
| Hæmoglobin | 76 per cent. |

(Haldane's modification of Gower's hæmoglobinometer). Blood from an œdematous foot contained :

| | | | | |
|-------------|---|---|-----------|---------------|
| Red cells | . | . | 4,250,000 | per cubic mm. |
| White cells | . | . | 6600 | „ „ „ |
| Hæmoglobin | . | . | 70 | per cent. |



After five minutes' vigorous massage of the foot blood withdrawn from it contained :

| | | | | |
|-------------|---|---|-----------|---------------|
| Red cells | . | . | 3,240,000 | per cubic mm. |
| White cells | . | . | 6000 | „ „ „ |

The last specimen of blood coagulated in 75 seconds at 22° C. (Wright's coagulometer)—*i.e.* considerably quicker than normal. Massage of normal tissues usually leads to a similar result to the one recorded above, but not to so great an extent. On pricking the skin of an area which has been quite recently massaged, tissue lymph pours out with the blood and dilutes it so that the number of

cells per cubic millimetre is decreased. The fact that the dilution in this case seems to have been greater than normal suggests that the œdema is due to tissue lymph and not to dilated lymphatics, because these would have been partially emptied by massage. The comparison of differential counts of white cells in the blood taken from the finger and the foot corroborates this view :

| | Finger. | Foot. |
|---------------------|----------------|----------------|
| Polynuclear . . . | 49.0 per cent. | 49.3 per cent. |
| Lymphocytes . . . | 40.3 „ | 40.7 „ |
| Hyaline cells . . . | 4.0 „ | 3.4 „ |
| Eosinophile . . . | 6.5 „ | 6.4 „ |
| Basophile . . . | 0.2 „ | 0.2 „ |

(Over 500 cells were classified.)

The similarity of these counts makes it evident that the diluting fluid either contained white cells in the same proportion as the blood or was free from white cells. The fact that the number of white cells was less in the blood taken from the foot than in that taken from the hand makes it certain that the diluting fluid was free from white cells—in other words, that it was tissue lymph and not lymph from lymphatics. The percentage of polynuclear cells is very low for a well-nourished child of five years. The eosinophilia is suggestive of a parasitic affection. *Filaria* were not seen in any of the blood-films ; this, of course, does not prove their absence. It is quite within the limits of possibility that the increase of eosinophile cells is due to some intestinal parasite and has no relation to the œdema. The examination of the blood of the other cases illustrates this.

| | T. R. (Case 1), aged 3 months. | J. H. (Case 3), aged 6 months. |
|-------------------|-----------------------------------|-----------------------------------|
| Polynuclear . . . | 32.4 per cent. | 29 per cent. |
| Lymphocytes . . . | 63.6 „ | 63 „ |
| Hyaline . . . | 1.6 „ | 6 „ |
| Eosinophile . . . | 2.4 „ | 2 „ |

I think that the above facts point towards the condition being due to œdema of the connective-tissue cells and not to dilated lymphatics.

CASE 3.—J. H—, a male child, aged 6 months, was admitted into the Belgrave Hospital under Mr. Jaffrey, whom I have to thank for his kindness in allowing me to describe the case. He presents a very similar condition of feet and legs to his sister (Case 2). The feet and legs are swollen, but the feet to a much greater extent than the legs. The skin of the legs is thickened. The skin around the joints

of the toes constricts them into three almost spherical masses; the œdema of the terminal phalanges is so great that the plane of the toes is tilted to almost a right angle to the long axis of the toes. The soles of the feet are affected as well as the dorsa; the heels appear to be normal. The measurement around the instep is 13.1 cm., about 1.3 cm. more than the same measurement of a child of the same age.

Since I have three cases only to add to the list it would be out of place for me to give a detailed summary of the numerous instances of this condition which have already been recorded. It may, however, be of interest to see how many varieties of this condition have been described and to note how they differ one from another. Hereditary angio-neurotic œdema was described by Osler in 1888, but this is quite a different disease from the one under discussion, for the œdema is not persistent and affects different parts at different times. It is, as is well known, associated with other neurotic symptoms, not rarely with intense abdominal neuralgia.

Milroy,* in 1892, wrote a paper on "An Undescribed Variety of Hereditary Œdema," in which he referred to twenty-two individuals in a family of ninety-seven affected with this complaint. The above cases I believe to be similar to those described by Milroy. Meige† recorded trophic œdema in two sisters; in one of these the disease was unilateral. These cases were not congenital. Debove‡ has termed a condition in which the œdema does not occur *en masse*, but which affects thighs or feet and not toes, as segmental œdema. In one of his cases the œdema began sharply at the gluteal fold and ceased at the popliteal space. There was not any thickening of the skin. Feindel§ portrays two sisters affected with œdema in which the disease was asymmetrical and progressive. Osler|| has described cases of scleroderma accompanied by œdema, but in these the scleroderma seemed to be the primary affection. Rolleston¶ described a family in which persistent œdema was a marked feature; in his cases the œdema was distinctly diminished by raising the legs; the skin was not thickened.

The conditions which must be recognised are: (1) Congenital hereditary œdema—(a) symmetrical, (b) asymmetrical; (2) trophic œdema developing in later life; (3) persistent hereditary postural œdema; (4) segmental œdema; (5) scleroderma with œdema.

* 'New York Medical Journal,' 1892.

† 'Nouvelle Iconographie de la Salpêtrière.'

‡ 'Presse Medicale,' 1902.

§ 'Gaz. Heb. de Med. et Chir.,' 1902.

|| 'Journal of Cutaneous and Genito-Urinary Diseases,' 1898.

¶ 'Lancet,' 1902.

